

Pediatric complex regional pain syndrome following maxillofacial trauma: A case report

Valeria Mitro¹, Giulio Cirignaco², Massimiliano Gilli¹, Francesco Giovacchini¹, Antonio Tullio³

From ¹Maxillofacial surgeon, Department of Maxillofacial Surgery, Azienda Ospedaliera di Perugia, Perugia, ²Maxillofacial surgery resident, Department of Maxillofacial Surgery, Università Degli Studi di Siena, Siena, ³Professor and Head, Department of Maxillofacial Surgery, Università degli studi di Perugia, Perugia, Italy

ABSTRACT

Complex regional pain syndrome (CRPS) has been described as a painful disorder with a different type of presentation and evolution months after the noxious event. The affected area shows color and vasomotor changes. Diagnosis of CRPS is hard and usually made by the Budapest criteria, as the pathological basis is still not completely known. We present this rare case that supports the idea of managing signs and symptoms with a multidisciplinary approach and supports the effective presence of a subtype of CRPS in children. This could make any other physician aware of possible diagnosis and, in the future, support the possible integration of this subtype in the Budapest criteria.

Key words: Children, Complex regional pain syndrome, Maxillo-facial, Multidisciplinary approach, Rare presentation

Complex regional pain syndrome (CRPS) has been described as a painful disorder with autonomic, sensory, and motor dysfunctions often disproportionate to the inciting event with an unknown etiology. The pain pathway from the peripheral to the central nervous system is overactive and sensitized to disturb a correct autonomic response [1]. The CRPS has variability in reports regarding its prevalence; it seems to affect more women than men with a male/female ratio of 1:2 and age ranging from 37 to 60 years. It is more common in the lower and upper extremities, with the latter affected twice than the former [2]. Diagnosis and treatment of CRPS are challenging because of its different types of presentation and evolution during months after the noxious event. The affected area can show vasomotor dysfunction along with swelling, edema, allodynia, and hyperalgesia, skin can appear erythematous, hyperemic, blue, purple, or pale [1,2]. This syndrome is commonly reported in the extremities, especially lower limbs and is directly linked with a trauma or injury that happened almost immediately or months before. It is also divided into CRPS I and II using the Budapest criteria, although they do not address pediatric CRPS [3,4]. A small number of cases of facial CRPS have been reported in adults after high-impact trauma, dental procedures, and maxillofacial surgery.

Here, we report a rare case of pediatric maxillo-facial CRPS that occurred in an 8-year-old girl after falling at home and was

successfully treated using a multidisciplinary approach. This report shows the effective presence of CRPS in children with an aim to support the integration of this subtype in the Budapest criteria.

CASE REPORT


A girl of 8 years old was referred to the maxillofacial surgery unit after a mild facial trauma that occurred in May 2022 after falling at home.

The general objective examination was negative. The facial objective examination showed edema and ecchymosis in the left zygomatic and periorbital regions. There was no facial muscle impairment, signs of bone fracture, or nerve injury. The patient was then discharged and enrolled in clinical follow-up, considering the low severity of clinical signs.

One month after the trauma, the patient was referred again to the maxillofacial unit with a clinical left-sided zygomatic swelling and persistent prickling neuropathic pain in the skin cheek. General examination revealed changes in the same region with a dry and blue skin color. Physical palpation demonstrated allodynia of the skin cheek and small hypoalgesia but no discomfort, eye movement, visual acuity, or facial muscle impairment (Fig. 1a). Different diagnoses included maxillo-facial fracture, infection, inflammatory, or vascular condition, and CRPS. The clinical symptoms did gradually improve during the next 30 days.

Correspondence to: Giulio Cirignaco, Department of Maxillofacial Surgery, Università degli studi di Siena, Siena, Italy. E-mail: giulioCirignaco@gmail.com

© 2023 Creative Commons Attribution-NonCommercial 4.0 International License (CC BY-NC-ND 4.0).

Access this article online	
Received - 13 July 2023 Initial Review - 25 July 2023 Accepted - 17 August 2023	Quick Response code 
DOI: 10.32677/ijcr.v9i9.4249	

After 8 months, the patient had a new upsurge in her clinical signs and symptoms with chronic hypoalgesia and important discomfort in the left malar skin region. We performed a computer tomography scan that did not show any complete or greenstick fracture of the zygomatic, maxillary, or orbital bone; moreover, different multispecialty consultation (ophthalmology, otorhinolaryngology, and neurology) was inconclusive. Routine blood tests and autoantibodies values were negative.

Since last year’s trauma, the child developed a psychological discomfort that did not lead to sleep her alone without parents, along with night cries and persistent pain. She was treated at the beginning with different corticosteroids and FANS without any improvement. To further confirm our diagnostic hypothesis of CRPS, we have adapted some questions to be understandable for our patient and used the Budapest criteria which are suitable for pediatric age (Fig. 2).

At this point, after 12 months of trauma, the patient began psychological therapy with the support of the physiotherapist and maxillofacial surgeon. Physical and occupational therapy was

performed 3 times a week with a rigorous home exercise program. Her therapy focused on function, using the affected area of the face and desensitization. After 3 months of the therapy, approximately 15 months after trauma, the symptoms started to improve. The parents reported a constant and gradual improvement in both psychological and physical aspects. At 1½ years, the patient no longer complains of pain, and skin discolorations are no longer evident (Fig. 1b).

DISCUSSION

CRPS is a condition of unknown etiology characterized by autonomic, sensory, and motor disturbances after a known trauma event with a different type of clinical presentation. The affected area can show a different type of skin color, edema, hyperalgesia, allodynia, warmth, and other peculiar clinical signs [1]. Paresthesia is rare and 75% of patients describe pain at rest and movement. Trophic skin color changes that occur over time are also associated with dysregulation of the autonomic somatosensory system [2].

The pathological basis of CRPS is yet to be completely known. Few hypotheses support neurogenic inflammation, vasomotor dysfunction [5], and a possible brain region connection that supports cortical activation on the application of orofacial stimulus [6]. In the literature, CRPS is divided into subtypes I and II and there is no direct and distinct correlation between the traumatic impact and the CRPS clinical symptoms. A noxious event is always the initiating mechanism, but in subtype I, the nerve lesion is non-demonstrable, whereas in II, it is demonstrable [2].

The diagnostic criteria of CRPS are called “Budapest criteria.” To make a diagnosis of CRPS, all the following statements are to be met: The patient has continuing pain disproportionate to any inciting event; the patient has one physical examination sign in the sensory, vasomotor, sudomotor/edema, and motor/trophic category, and the patient report 1 symptom in 3 of the categories or 1 sign in the 2 categories below. No other diagnosis better explains the patient’s presentation.



Figure 1: Three-quarter view of the patient showing (a) edema of the left malar region and skin discoloration; (b) resolution of pathological signs resolution

1) Ti ricordi di preciso cosa è successo quando sei caduta?

2) Hai sentito tanto dolore subito dopo essere caduta? Scegli la foto che ti rappresenta

0 Non fa per niente male 2 Fa male solo un pochino 4 Fa male un po' di più 6 Fa ancora più male 8 Fa tanto male 10 Fa un male incredibile

3) Il dolore è cambiato nei giorni successivi dopo essere caduta o è sempre rimasto costante? Scegli la foto che ti rappresenta

0 Non fa per niente male 2 Fa male solo un pochino 4 Fa male un po' di più 6 Fa ancora più male 8 Fa tanto male 10 Fa un male incredibile

4) Sentì ancora dolore oggi? Scegli la foto che ti rappresenta

0 Non fa per niente male 2 Fa male solo un pochino 4 Fa male un po' di più 6 Fa ancora più male 8 Fa tanto male 10 Fa un male incredibile

5) Percepisci tutto nella zona dove sei caduta o ogni tanto pensi di sentire diverso? Toccando con le dita cambia?

6) Quante volte ti è capitato di vedere la pelle avere un colore diverso sulla guancia a sinistra? Se si quante volte è successo?

7) Ti è mai capitato di vedere gonfio? Se si quante volte è successo?

Figure 2: Budapest criteria containing several questions

Very few articles are cited for CRPS of the orofacial region [2,7,8]. All these cited cases describe a noxious event in children, major features of CRPS, and a particular multidisciplinary approach as in children, psychological issues play a more prominent role, and neurological symptoms are less pronounced [6]. The diagnosis of CRPS in children should be considered after excluding other pathologies as it is a rare event and Budapest criteria does not include them.

The chosen approach was not to undergo invasive tests such as skin biopsy or electroneurography used in the other case reports cited above but to understand an eventual connection between the traumatic events and clinical presentation with the idea of a possible CRPS considering the age and the evolution of symptoms. Different treatments are described for CRPS, starting from pharmacological to surgical [9], even if there are no known prospective randomized clinical trials of medication for pediatric cases [4]. Our experience supports the idea of managing a multidisciplinary approach made by a surgeon, physician, physiotherapist, psychologist, and, if necessary, pain therapist. In fact, in our experience, the patient gradually improved after undergoing behavioral and psychological support. Her parents told us that the night cry and sharp pain recrudescence disappeared. The psychological approach has been demonstrated to be effective when tailored to the individual personality to make the patient understand and accept the different grades of possible pain management methods.

CONCLUSION

Our report shows that a multidisciplinary approach has a crucial role in treating this type of patient; moreover, our case presentation supports the effective presence of a subtype of CRPS in children. This could make any other physician aware of the possible diagnosis and, in the future, support the possible integration of this subtype in the Budapest criteria.

ACKNOWLEDGMENTS

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation and with the Helsinki Declaration of 1964. Informed consent was obtained from all patients for being included in the study.

REFERENCES

1. Neumeister MW, Romanelli MR. Complex regional pain syndrome. *Clin Plast Surg* 2020;47:305-10.
2. Heir GM, Nasri-Heir C, Thomas D, Puchimada BP, Khan J, Eliav E, *et al.* Complex regional pain syndrome following trigeminal nerve injury: Report of 2 cases. *Oral Surg Oral Med Oral Pathol Oral Radiol* 2012;114:733-9.
3. Harden NR, Bruehl S, Perez RS, Birklein F, Marinus J, Maihofner C, *et al.* Validation of proposed diagnostic criteria (the "Budapest Criteria") for complex regional pain syndrome. *Pain* 2010;150:268-74.
4. Katholi BR, Daghestani SS, Banez GA, Brady KK. Noninvasive treatments for pediatric complex regional pain syndrome: A focused review. *PM R* 2014;6:926-33.
5. Marinus J, Moseley GL, Birklein F, Baron R, Maihofner C, Kingery WS, *et al.* Clinical features and pathophysiology of complex regional pain syndrome. *Lancet Neurol* 2011;10:637-48.
6. Lee YH, Lee KM, Kim HG, Kang SK, Auh QS, Hong JP, *et al.* Orofacial complex regional pain syndrome: Pathophysiologic mechanisms and functional MRI. *Oral Surg Oral Med Oral Pathol Oral Radiol* 2017;124:e164-70.
7. Goenka A, Aziz M, Riley P, Vassallo G. Complex regional pain syndrome involving the face following snowball injury. *Eur J Pediatr* 2014;173:397-400.
8. González-Cantero Á, Sánchez-Moya AI, Pérez-Hortet C, Martínez-Lorenzo E, Gómez-Dorado B, Molina-Figuera E, *et al.* Complex regional pain syndrome of the face in a child. *Int J Dermatol* 2018;57:1502-3.
9. Melf-Marzi A, Böhringer B, Wiehle M, Hausteiner-Wiehle C. Modern principles of diagnosis and treatment in complex regional pain syndrome. *Dtsch Arztebl Int* 2022;119:879-86.

Funding: Nil; Conflicts of interest: Nil.

How to cite this article: Mitro V, Cirignaco G, Gilli M, Giovacchini, F, Tullio A. Pediatric complex regional pain syndrome following maxillofacial trauma: A case report. *Indian J Case Reports.* 2023;9(9):286-288.